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Lemierre syndrome: the forgotten disease—a case series

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Abstract

Background The aim of this work is to share the authors' experience and assist therapeutic decision-making in the management of Lemierre syndrome (LS). This is a retrospective descriptive study carried out in the stomatology, maxillofacial surgery, and ENT departments of the teaching hospital of Treichville (Abidjan, Côte d'Ivoire). LS cases managed from 2013 to 2023 were included in the study.

Results Eight patients were enrolled (six men and two women). Internal jugular vein thrombosis was unilateral in 7 cases and bilateral in one. No germs were found in four patients. In one patient, a bacterial co-infection was responsible for the thrombosis. Surgical debridement was systematically performed in cases of necrosis. Anticoagulation was justified in three patients. No deaths were observed.

Conclusion This work describes the epidemiological, clinical, therapeutic, and evolutionary aspects of LS. The management of LS should not be improvised. The choice of surgical debridement and anticoagulation depends on specific situations.

Keywords Lemierre syndrome, Thrombophlebitis, Anticoagulation, *Fusobacterium necrophorum*

Mots-clés Syndrome de Lemierre, Thrombophlébite, Anticoagulation, *Fusobacterium necrophorum*

Background

Lemierre syndrome (LS) is a septic thrombophlebitis of the internal jugular vein (IJV). It usually complicates an oropharyngeal or cervico-facial infection. The common germ is *Fusobacterium necrophorum* and other germs are exceptional [1]. It is a rare, serious, and life-threatening condition if the diagnosis is late and/or the management is inadequate. In tropical Africa, very few studies report cases of LS. However, it is certainly underdiagnosed and its incidence is probably underestimated [1, 2]. The authors report their experience in the management of LS through a series of eight cases from 2013 to 2023. They describe epidemiological, clinical, therapeutic, and evolutionary aspects.

Methods

This is a retrospective and descriptive study, carried out in the Maxillo-Facial Surgery and ENT departments of the Teaching Hospital of Treichville in Abidjan (Côte

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Table 1 Epidemiological, clinical, therapeutic, and evolutionary aspects of the patients

N°	Unit of management	Age (Years) and sex	Comorbidities and factors favoring	Aetiology	Vessel and side of thrombosis	Isolated germs	Antibiotic received	Anticoagulation	Resection of the IJV	Recanalization of the IJV after treatment	Evolution
1	MFS	45, M	Anti-inflammatory medication	Angina + necrotizing cellulitis	Left IJV	None	Ceftriaxone + metronidazole	Enoxaparin	Yes	Not applicable	Healing
2	MFS	52, F	Anti-inflammatory medication	Right tonsil foreign body (fishbone)	Right IJV + superior vena cava	<i>Staphylococcus aureus</i>	Ceftriaxone + gentamicin	Enoxaparin then acenocoumarol	No	Yes	Healing
3	MFS	34, M	Anti-inflammatory medication	Odontogenic cellulitis	Left IJV	<i>Staphylococcus aureus</i>	Ceftriaxone + gentamicin + metronidazole	No	No	Yes	Healing
4	MFS	19, M	Tuberculosis	Tuberculous cervical lymphadenopathy	Right IJV	<i>Mycobacterium tuberculosis</i> et <i>Levinea sp</i>	Rifamycine Isoniazid Pyrazinamide Ethambutol (RHZE) + Ceftaxime + Imipenem	No	No	Yes	Healing
5	MFS	28, F	Anti-inflammatory medication	Necrotizing angina	Right IJV	<i>Escherichia coli</i>	Ceftriaxone + metronidazole	Enoxaparin	No	Yes	Healing
6	MFS	69, M	Not found	Retropharyngeal abscess	Right IJV + left IJV	None	Ceftriaxone + metronidazole	Enoxaparin	No	Yes	Healing
7	MFS	23, M	Anti-inflammatory medication	Odontogenic cellulitis	Right IJV	None	Ceftriaxone + metronidazole	No	No	Yes	Retropharyngeal abscess then gone without medical advice
8	ENT	30, M	HIV Infection + Anti-inflammatory medication	Angina + cervical abscess	Right IJV	None	Ceftriaxone + metronidazole	No	No	Yes	Healing

M male, F female, MFS maxillofacial surgery, ENT ear, nose, and throat, HIV human immunodeficiency virus

d'Ivoire). We collected all patients' files hospitalized in the above services, from 2013 to 2023, in whom the diagnosis of LS was made. The diagnosis of LS was confirmed by a cervico-facial CT scan with contrast injection at the admission in all patients. The CT scan was also performed after the treatment (at least 3 months after treatment) to attest to the complete drainage of the pus and the recanalization of the IJV. In all patients, a cardiopulmonary auscultation, an electrocardiogram, and a chest X-ray were realized. Blood examinations including blood cell count, venous glycemia, urea, creatinine, C-reactive protein, blood culture, and retroviral serology were also realized. The presence of pus and/or necrosis systematically motivated a surgical debridement or the surgical drainage of the pus. Pus collected after surgical debridement or drainage was directly examined and cultured in aerobic and anaerobic media to identify the germs involved. The parameters studied were age, sex, isolated germs, comorbidities, treatment received, the recanalization of the IJV, and the outcome of the treatment. The disappearance of the infectious syndrome, the total drying up of the suppuration, and the absence of extension of the necrosis were considered as criteria of healing.

Results

Six men and two women were included in the study (sex ratio=3). The average age was 37.5 years. Table 1 summarizes the epidemiological, clinical, therapeutic, and evolutionary aspects of the patients. Seven patients were followed up in the Maxillofacial Surgery department and one patient in the ENT department. Taking anti-inflammatory medication was the most frequent situation associated with cervical infection (Table 1). Cervical and/or oropharyngeal ulceration or necrosis was observed in four patients (Figs. 1 and 2) and IJV thrombosis was always homolateral to the necrosis except for one patient (Patient 6). Resection of the IJV was performed in one patient (Patient 1) due to the thrombus which occupied all the length of the IJV, the persistence of the infectious syndrome, and the extension of cutaneous necrosis despite medical treatment (Fig. 3).

Fusobacterium necrophorum was not isolated in any patient. Four other germs were identified in the samples, including two in the same patient (Table 1). In four cases, no germs were isolated. The right IJV was thrombosed in 5 cases versus 2 cases for the left IJV. In one patient (Patient 6), both IJV were thrombosed (Fig. 2). In all patients, we observed recanalization of the IJV when it had not been resected. The association Ceftriaxone + Metronidazole was always the first empiric antibiotic combination administered. Antibiotic therapy was secondarily adapted to the results of the antibiogram when a germ was identified.



Fig. 1 Patient 2. **A** Right cervical necrosis. **B** Cervical CT scan in axial section after contrast injection: Right internal jugular vein thrombosis (arrowhead). **C** Thoracic CT scan in axial section: superior vena cava thrombophlebitis (arrow)

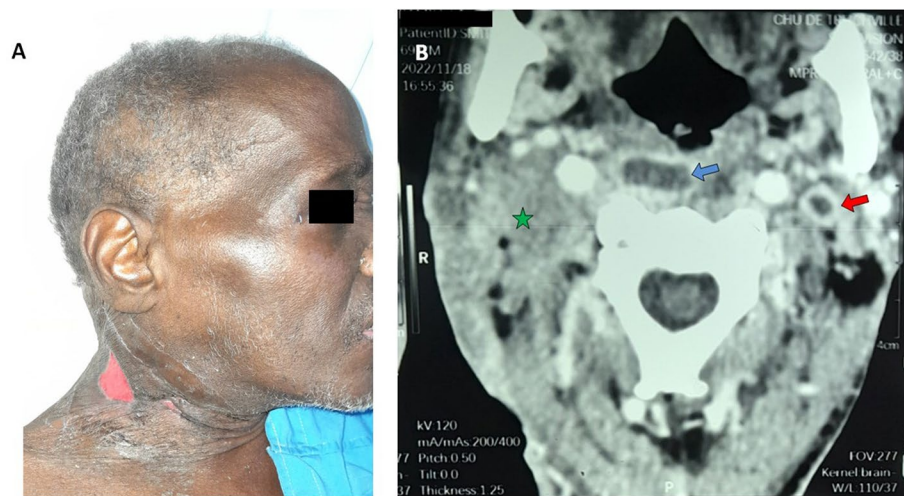


Fig. 2 Patient 6. **A** Right cervical skin loss of substance. **B** Cervical CT scan (axial section) with contrast injection showing bilateral internal jugular vein thrombosis (red arrow, thrombosis of the left internal jugular vein; blue arrow, retropharyngeal abscess; green star, right internal jugular vein non-visible due to thrombosis)

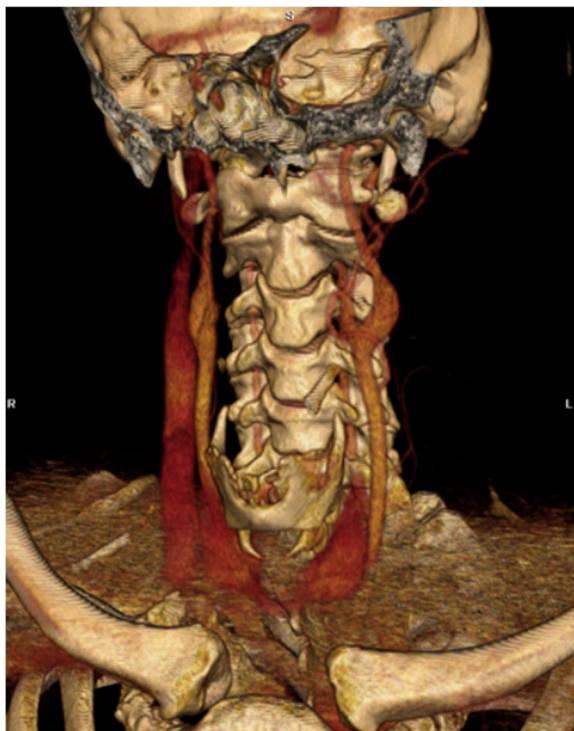


Fig. 3 Patient 1. Three-dimensional reconstruction of the cervical vessels after resection of the left internal jugular vein (not visible)

Discussion

LS or “forgotten disease” is a rare disease. Its annual incidence is estimated at 1 case/1,000,000 habitants per year [2, 3]. There is no formal consensus on its management. However, when there is necrosis, it systematically

requires a surgical debridement. The debridement is an essential part of the management because it eliminates necrosis favoring anaerobic conditions which increase sepsis [1, 4]. In all patients, surgical debridement was performed and improved prognosis. No deaths were observed.

In patient 6, bilateral IJV thrombosis was observed. It is a very rare situation which is poorly described in the literature [5]. Although identified as the common germ in LS, *Fusobacterium necrophorum* was not found in any of the patients in this series. Its culture and isolation are difficult, but the absence of germ isolation should not, however, lead to a denial of diagnosis [4]. In patient 4, two germs were identified, including *Mycobacterium tuberculosis*. To our knowledge, this is the only case of LS reported in the Anglo-Saxon literature caused by a co-infection. Two mechanisms may explain the occurrence of thrombosis of IJV in case of tuberculous cervical lymphadenopathy. It is either the compression of the IJV by lymphadenopathy which partially or totally interrupts the vascular flow or the vascular inflammation favorable to the Virchow triad (stasis, hypercoagulability, and alteration of the endothelium) and consecutive to the lymphatic infection of contiguity [6]. Several families of antibiotics can be used for treatment. In all cases, in the beginning, antibiotic therapy is empiric, active on anaerobic germs (especially on *Fusobacterium necrophorum*), broad spectrum, and by injection. The use of Metronidazole is recommended and has been proven effective in LS [7]. The resection of IJV in LS management is poorly described in the literature and remains a last resort. It is sometimes the only way to stop the progression of the

thrombosis and the infectious process thus avoiding the occurrence of pulmonary septic emboli [4]. It was performed in one of the patients (Patient 1) due to the total occlusion of IJV, the persistence of infectious syndrome, and the extension of cervical necrosis despite medical treatment (antibiotics and anticoagulation). The evolution was quickly favorable after the resection of the IJV with the disappearance of the infectious syndrome.

The use of anticoagulation in the treatment of LS is highly controversial and the efficacy discussed [1, 7]. In this series of cases, the use of anticoagulation was justified in only three cases. For patient 1, anticoagulation was administered because the occlusion of the IJV was complete along its entire length with a high risk of embol migration to the lungs. In patient 3, the extension of the thrombus to the superior vena cava could lead to migration to the right heart and the dissemination of septic emboli to the lungs. In patient 6, anticoagulants were administered primarily to prevent venous thromboembolic disease in an elderly and bedridden patient.

In all the patients, the diagnosis of Lemierre syndrome was not made fortuitously. The presence of a cervical tumefaction or cervical necrosis systematically motivated the search for a possible thrombosis of the IJV by the realization of a cervical CT scan.

We consider that any acute, infectious, and painful cervical swelling, associated or not with cervical necrosis or following a pharyngeal infection, must be investigated with a cervical CT scan, the key examination for the diagnosis of LS [1, 4, 8].

Conclusion

The management of LS must not be based on any therapeutic improvisation. Anticoagulation is not systematic but its use may be justified in certain specific situations. Surgical debridement is mandatory in case of necrosis. Empiric antibiotic therapy must be immediately active on anaerobic bacteria, more particularly *Fusobacterium necrophorum*. Proper management improves the prognosis.

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Authors' contributions

All authors had full access to all of the data in the study and take responsibility for the integrity of the data. AS was responsible for the concept and design, acquisition and interpretation of the data, drafting of the manuscript, and critical revision of the manuscript. CA was responsible for the concept and design, interpretation of the data, critical revision of the manuscript, and supervision. DN and IG were responsible for the interpretation of the data and critical revision of the manuscript. DM and AREY were responsible for the critical revision and corrections. EK was responsible for the concept and supervision. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate

Ethical approval for the current study protocol was obtained from the Research Ethics Committee of the Teaching Hospital of Treichville (N°027/23). Informed written consent to participate in the study was provided by all participants.

Consent for publication

Written consent for publication is taken from all participants.

Competing interests

The authors declare that they have no competing interests.

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